CASE REPORT

CE 2.0 Contact Hours

Frontal Fibrosing Alopecia

Kate Kimes, Kim Carlson

ABSTRACT: Frontal fibrosing alopecia (FFA) is a form of hair loss with an unknown etiology seen in postmenopausal women. Its clinical presentation is characterized by alopecia along the frontal hairline that extends posteriorly with scarring. On examination, perifollicular hyperkeratosis and erythema may be present. Histologically, FFA is identical to lichen planopilaris. Unfortunately, there is no consistently effective treatment. Various treatment options may be tried, and eventually, patients may opt to use camouflage such as wigs to disguise the alopecia. In time, most cases will stabilize, with or without treatment. More research is needed not only to determine etiology but also to establish evidence-based treatment options for FFA.

Key words: Frontal Fibrosing Alopecia, Lichen Planopilaris, Bioidentical Hormones, Autoimmune Alopecia

CASE REPORT

History of Present Illness

A 66-year-old Caucasian woman, P. B., presented to the dermatology clinic as an established patient who was last seen 6 months prior. The chief complaint at this visit was a "red and scaly rash" on her frontal scalp that had been there for months. P. B. did not complain of hair loss until questioned by the nurse practitioner if she had noticed any change in her hairline over the years. P. B. then admitted that frontal scalp hair had been shedding for 1–1.5 years, with loss on the eyebrows, arms, and legs progressing since starting about 4 years prior. P. B. stated, "My tennis visor sits in that area," and she admitted to playing tennis three to four times a week with her hair in a ponytail. P. B. denied any associated symptoms of itching, pain, or burning.

P. B. had a history of nonmelanoma skin cancer, and a previous biopsy of a lesion on the proximal dorsal nose revealed "acanthosis with purulent scale crust, transected, which may represent changes overlying a folliculitis. Underlying neoplasm cannot be detected." She denied a past history of alopecia, although she had a prescription of desonide 0.05% cream from an office visit with a different provider for the symptom of "dryness" on the face. The examination on that date revealed "erythematous papules" on the face without a formal diagnosis. The patient denied any current oral medications or supplements other than bioidentical hormone pellets, which were started 1–1.5 years prior. P. B. denied other pertinent past medical history including autoimmune or endocrine disorders.

Physical Examination

Examination showed a regressing hairline at the anterior frontal scalp with a few hairs maintained in previous hairline (Figure 1). There was perifollicular erythema, mild hyperkeratosis, and decreased follicular openings (Figure 2).

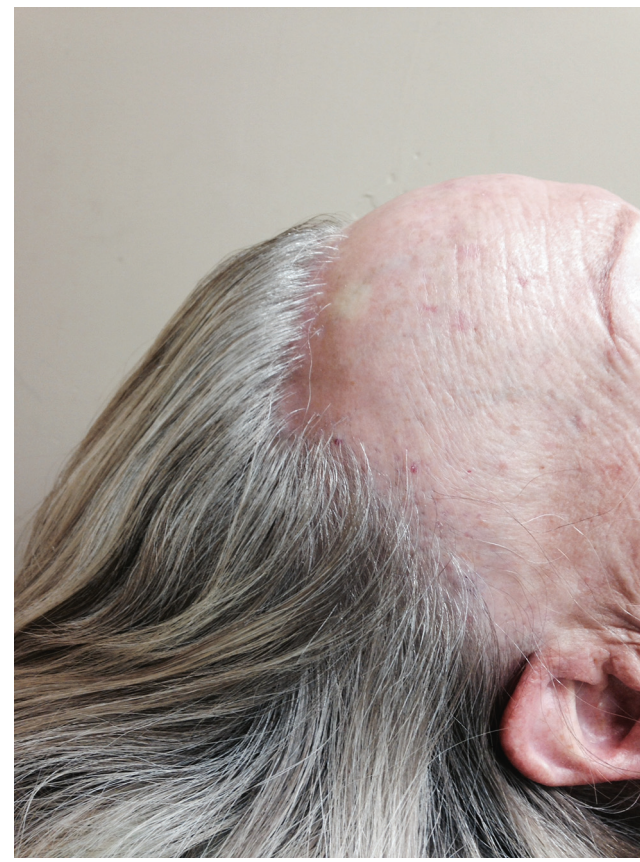

FIGURE 1. Alopecia along the frontal hairline.

Kate Kimes, BA, *Arizona College of Osteopathic Medicine, Midwestern University, Glendale, AZ.*

Kim Carlson, RN, MSN, NP-C, DCNP, *MacKenzie Dermatology, Prescott, AZ.*

The authors declare no conflict of interest.

Correspondence concerning this article should be addressed to Kate Kimes, BA, Arizona College of Osteopathic Medicine, Midwestern University, 7101 W. Beardsley Rd. #421, Glendale, AZ 85308. E-mail: katekimes@gmail.com

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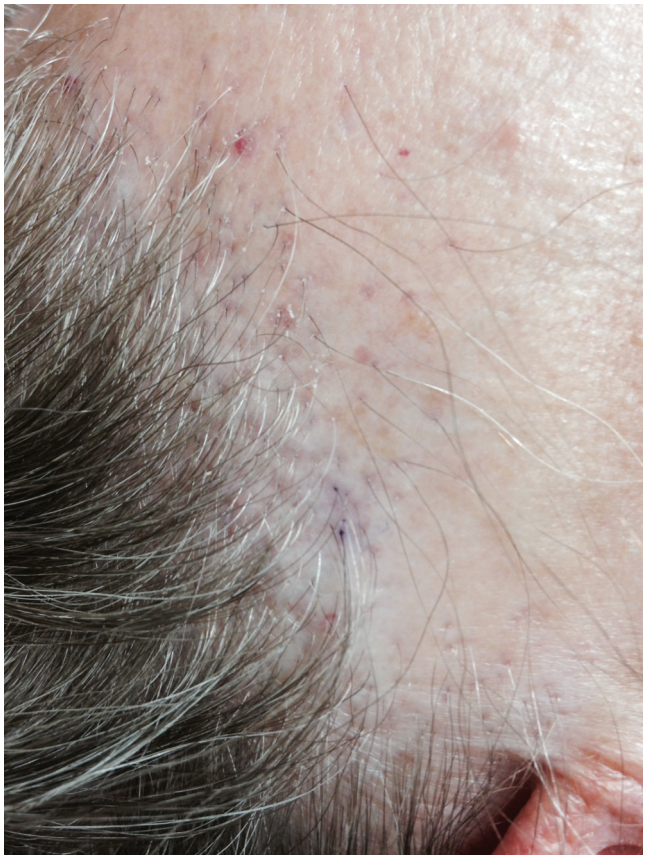

FIGURE 2. Perifollicular erythema, hyperkeratosis, and scarring present.

No vellus hairs or broken hairs that are narrower toward the scalp (known as exclamation hairs) were present. No pustules, papules, excoriations, or pigmented casts were present. A gentle hair pull test at the anterior hairline and midcrown was negative. No other focal areas of alopecia were evident on the scalp, but there was diffuse hair loss from the arms, legs, and eyebrows. Her eyelashes were intact and of expected size and number. There was no nail pitting, pterygium, oral Wickham striae, or ulcerations (Mubki, Rudnicka, Olszewska, & Shapiro, 2014).

Pathology

Histologic features of two 4-mm punch biopsies in formalin from the left lateral forehead at the hairline and mid-anterior crown showed "scarring alopecia, favor lichen planopilaris" with a comment, "The presence of scattered eosinophils makes lupus less likely."

DISCUSSION

Frontal fibrosing alopecia (FFA) is a form of hair loss most often seen in postmenopausal Caucasian women. In a recent multicenter review of 355 patients with FFA, 98.5% were Caucasian, 97% were women, and 86% of the women were postmenopausal (Vañó-Galván et al., 2014). This has led some to suggest an androgen-driven cause hypothesis, but currently, the etiology is unknown. As the name suggests, it is a scarring alopecia. It is considered by most to be a variant of lichen planopilaris (LPP), a perifollicular inflammatory disease. The inflammation is characterized by a cell-mediated immune response that may be considered an autoimmune disease, as it is often associated with other autoimmune diseases such as thyroid abnormalities (Habif, 2010; MacDonald, Clarke, & Holmes, 2012).

The clinical presentation of FFA is characterized by alopecia along the frontal hairline that extends posteriorly with scarring. Patients may complain of an associated pain, stinging, or burning in the affected areas. On examination, perifollicular papules, erythema, and scaling may be present (Habif, 2010). The alopecia may or may not involve the eyebrows, eyelashes, and/or body hair. In a 2012 study of 60 patients with FFA, 73% had eyebrow loss, 25% had loss of body hair, and only 3% had lost eyelashes (MacDonald et al., 2012).

The histology of FFA is the same as LPP: In early stages, there is a lichenoid interface inflammation at the hair follicles and within the perifollicular dermis (Habif, 2010). Later, a perifollicular fibrosis is present (MacDonald et al., 2012). A 2010 histopathological study of 13 patients with FFA showed that the histology of areas of alopecia on the body showed the same features as on the scalp (Chew, Bashir, Wain, Fenton, & Stefanato, 2010).

Treatment Options

Unfortunately, there is no consistently effective treatment for FFA. First-line therapy is often corticosteroids either topically, intralesionally, or orally (Habif, 2010). Other medications and medication combinations may be tried such as hydroxychloroquine, topical calcineurin inhibitors, or minoxidil; however, each has inconsistent results (MacDonald et al., 2012). In patients also presenting with androgenic alopecia in one study, finasteride with minoxidil was attempted with no significant improvement (Rallis, Gregoriou, Christofidou, & Rigopoulos, 2010). However, a systematic review found that oral 5-alpha reductase inhibitors had a good clinical response in 45% of patients and were the most effective treatment (Rácz, Gho, Moorman, Noordhoek Hegt, & Neumann, 2013). Currently, it is not obvious whether treatment affects long-term outcomes. A 2009 British study found that, in most cases, the disease stabilized over time regardless of treatment. However, the rate of progression of the disease and the amount of time before the disease was stable were variable (Tan & Messenger, 2009). Many patients eventually focus on camouflage of the disease with wigs (MacDonald et al., 2012).

FFA is an interesting, albeit frustrating, disease for the clinician. Although it can be diagnosed clinically by its location and by its comparable histology with LPP, the origin remains a mystery. The appearance is distressing to patients, and regrettably, there are no reliably effective treatments. Various treatment options may be tried, and eventually, patients may opt to use camouflage such as wigs to disguise the alopecia. In time, most cases will stabilize, with or without

treatment. In this presentation of FFA, concomitant start of bioidentical hormones is an intriguing aspect. A review of the literature did not reveal any previous examples of this association at this time. More research is needed not only to determine etiology and treatment for FFA but also to investigate if bioidentical hormone replacement is a factor in progression or a potential therapeutic treatment option.

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